CLAIMS

A method of treating neurodegenerative disease in a mammal comprising the steps of introducing a therapeutic effective amount of a chaperone or chaperone-like-compound into the neurological system of the mammal.

The method of claim 1, wherein the introducing step includes introducing the chaperone or chaperone-like-compound into the mammal by gene therapy

SUR BZ)

10

15

20

25

30

The method of claim 1, wherein the introducing step includes directly injecting the chaperone or chaperone-like-compound into the mammal.

A method for screening for a test compound for chaperone-like activity for the treatment of neurodegenerative diseases comprising the steps of:

introducing the test compound into transfected cells in tissue culture, wherein such transfected cells produce protein aggregate; and

measuring the quantity of protein aggregate, wherein a test compound which decreases the quantity of protein aggregate as compared to control cells has chaperone activity.

5. A method for screening for a test compound for chaperone-like activity for the treatment of neurodegenerative diseases comprising the steps of:

introducing the test compound into an animal which models neurodegenerative disease;

5

10

15

20

25

30

allowing said animal to develop; and subsequently measuring the quantity of aggregates in said animal wherein decreased aggregate formation over control animals indicates chaperone-like activity.

- 6. A method of treating neurodegenerative disease in a mammal comprising the step of introducing a therapeutically effective amount of a compound into said mammal wherein said compound increases the effective concentration of a chaperone in the neurological system.
- 7. A method of treating neurodegenerative disease in a mammal comprising the step of introducing a therapeutically effective amount of a compound into said mammal wherein said compound increases the effective concentration or enhances the activity of a proteasome in the neurological system.
- 8. A method for screening for a test compound which increases proteasome activity for the treatment of neurodegenerative diseases comprising the steps of:

introducing the test compound into transfected cells in tissue culture, wherein such transfected cells produce protein aggregate; and

measuring the quantity of protein aggregate, wherein a test compound which decreases the quantity of protein aggregate is selected.

9. A method for screening for a test compound which increases proteasome activity for the treatment of neurodegenerative diseases comprising the steps of: 5

introducing the test compound into an animal which models neurodegenerative disease;

allowing said animal to develop; and subsequently measuring the quantity of aggregates in said animal wherein a compound which shows decreased aggregate formation over control animals is selected.

10. Transgenic mice capable of overexpression of HDJ-2.

ALA

ADDB3